THE SYNDROME OF COAGULATION MASSIVE ET XANTHOCHROMIE OCCURRING IN A CASE OF TUBERCULOSIS OF THE CERVICAL SPINE.

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THE following case occurred on the orthopedic service of Dr. Lovett at the Children's Hospital. Through his kindness it is possible for me to report this illustration of a symptom-complex which has been little described in American literature. Its study may serve to shed some light in the future upon similar cases, seemingly obscure, in which an equally characteristic spinal fluid may be discovered.

CASE REPORT.—R. B., male, aged three years; birthplace, Boston; hospital record, 5786.

Family History. Father, mother, and three other children living and well. Paternal uncle died of tuberculosis of the lungs. No children dead. Mother has had no miscarriages.

Past History. Third child, full term, easy labor. Breast-fed for eighteen months. Eczema at three months; cholera infantum at eighteen months; bronchopneumonia during winter. Has always been a weak, inactive child.

Present Illness. Began September, 1914. Parents noticed that the child commenced to hold his head in left-sided torticollis. The latter getting increasingly worse, has been accompanied by some feverishness and considerable irritability. Appetite good; bowels regular.

Physical Examination. A poorly-developed and nourished child, sick and feverish; head, otherwise normal, held in marked torticollis. Eyes: pupils equal and react to light and distance. No nasal discharge. Throat clear. Teeth in good condition. Chest: fair expansion; no abnormal changes in breath sounds nor in percussion note. Heart: area is normal; sounds of good rhythm, clear, and loud. Abdomen normal, no masses.

Local History. Patient stands with a marked left cervicodorsal, right dorsal, and left dorsolumbar scoliosis, with the head tilted to the left and the chin rotated a little to the right. Walks with a very unsteady gait. Lying in bed no facial asymmetry is noted. Considerable elevation of the right shoulder and the inferior angle of the left sternocleidomastoid muscle, especially at the sternal attachment; no contraction of the trapezius. Head held in approximately 60 degress of left lateral flexion, 40 degrees of anterior flexion, and 30 degrees of rotation to the right. All joint motions elsewhere throughout the body are free. General signs of old rickets, rather square head, enlarged epiphyses at elbows and knees, prominent abdomen, some outward bowing of the femora and tibiæ, slight Harrison's groove, and rachitic rosary.

Reflexes: Knee-jerks present on both sides. No clonus; no Babinski; no Oppenheim.

Von Pirquet reaction, forty-eight hours after admission, human negative, bovine negative.

Urine on admission: reaction acid; no albumin; no sugar; no acetone. Microscope shows urates and few blood cells. Blood on admission: leukocytes, 16,300; polymorphonuclears, 57 per cent; lymphocytes, 43 per cent.

December 3, 1914. Position of the head has gradually corrected itself since admission. Much less tenderness on moving the head. Guards the neck very carefully and will not voluntarily move the head. Temperature has been running a little evening rise, otherwise his general condition has remained fairly good.

December 15. Patient moves his head to the right on suggestion, but still prefers to hold it to the left. In relation to possible intestinal origin of the trouble, child placed on a buttermilk diet. Also taking aspirin.

December 22. Child now holds his head without lateral flexion; can turn with fair degree of ease both to right and left. The right trapezius is held rigid. Aspirin and buttermilk diet discontinued because of digestive upset.

December 24. Marked rigidity of the neck posteriorly. No headache; no Kernig sign present. A lumbar puncture was made; under slightly increased pressure, 10 c.c. of clear yellow fluid with a slight greenish tinge was obtained. Cell count, three per field. Lymphocytes, 88 per cent.; globulin reaction (ammonium sulphate) positive. Upon standing two hours, moderate coagulation. No organisms on smear or blood culture. The removal of the fluid seemed to alleviate the retraction of the neck.

December 28. A second lumbar puncture, not under pressure, shows the same distinctly yellow color. Renewed examination of the child shows a slight remaining left torticollis and dilatation of the left pupil; marked apathy, insensible to the prick of a pin; any real anesthesia impossible to determine. Right side of the body, from the face down, shows weakness. Patient unable to raise the leg, and able only with difficulty to lift the arm. Knee-jerks exaggerated; ankle-clonus present.

December 30. Examination by Dr. J. J. Thomas: Cervical sympathetic paralysis on right side; weakness of sternomastoid; contraction of pupil; narrow palpebral opening and dryness of right side of face. Right arm and chest give suggestion of beginning flaccid paralysis; the right leg gives a suggestion of beginning spastic paralysis with ankle-clonus. Positive Babinski; exaggerated knee-jerks. No definite anesthesia can be made out.

January 2, 1915. Examination by Dr. Harvey Cushing: Face shows peculiar sardonic smile. The right eye shows narrowing of the palpebral fissure, some inophthalmos, and a contracted pupil. Vasodilatation persists on pinching the right cheek. Diagnosis of right sympathetic paralysis, cervical. Both pupils react to light; ocular movements normal; globes parallel. Ophthalmoscopic examination negative on the left side and impossible on the right side because of contracted pupil. Tongue protrudes in the median line. Body shows peculiar insensibility to pin-prick. There does not appear to be any complete anesthesia. Patient unable to move the right leg; only in part the right arm; deep reflexes easily elicited without clonus; perhaps better on right than on left. Babinski sign positive, both right and left. No abdominal reflex obtained. Positive findings: (1) muscular weakness on right side of body; (2) right cervical sympathetic paralysis.

January 11. New Roentgen-rays of the cervical vertebræ show an evident carious development in the bodies of the third and fourth vertebræ, a condition not noted in earlier radiographs. General condition of the patient shows slow downward progress. Head traction applied. Wassermann reaction negative.

January 18. General condition very poor. Lungs seem to show pulmonary edema; numerous moist rales throughout; no definite areas of impairment. Breath sounds exaggerated but not bronchial. Face shows on the right side marked flushing, plainly demarcated at the midline; left side shows pallor. This is evidently due to the paralysis above noted.

January 25. Despite the frequency and persistency in the pulmonary edema the patient's condition remains about the same, and the child has been, for the most part, very comfortable. He is kept in a sitting posture, which seems to afford some relief.

January 28. Condition today the same until 10 P.M., when breathing suddenly ceased.

DISCUSSION. In this case, the two interesting points are the cervical sympathetic paralysis and the results of the examination of the spinal fluid.

The records of the cases of tuberculosis of the cervical spine admitted to the orthopedic house service of the Children's Hospital from August 1, 1907, to February 1, 1915, reveal the fact that in a total of twenty-three not one shows the paralysis above noted. Of several standard text-books on orthopedic surgery, only that of Tubby[1] mentions the possibility of its occurrence.

This paralysis may result from torticollis *per se*, without changes in the spinal cord.[2] In this case, however, the radiographic demonstration of lesions in the fourth cervical body supports the assumption of a spinal origin. The syndrome is the result of interference with fibers in the cervical cord running from the ciliospinal centre.

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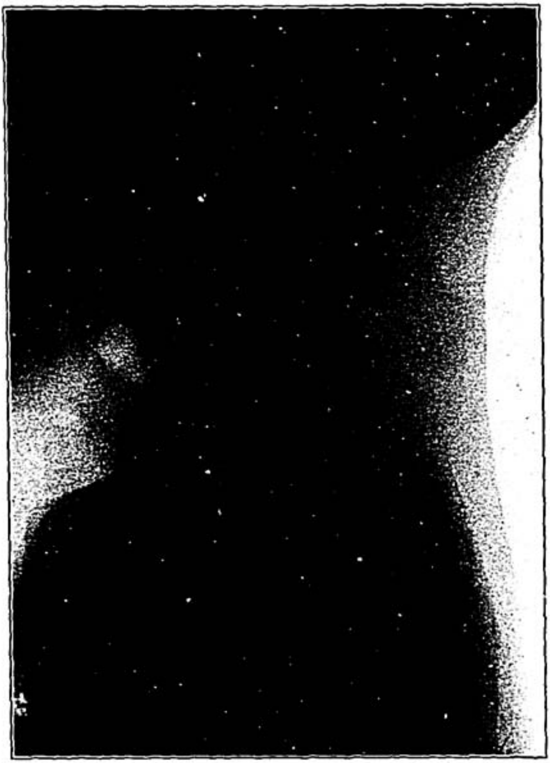
[1] Deformities Including Diseases of Bones.

[2] See Tubby, loc. cit.



Roentgenogram of the cervical vertebræ, showing caries in the bodies of the second and fourth vertebræ.

A review of the work of W. Mestrezat,[3] in which the characteristics of the spinal fluid in Pott's disease have been well summarized, will lend differential interest to the consideration of the present case.

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[3] Le Lequide Cephalo-Rachidien, normal et pathologique. Paris. 1912.

As quoted by Mestrezat, Foi and Sicard found when the disease was at its height the following:

1. "Xanthochromie," or distinct yellow color, quite pronounced.
2. An abundant coagulation, either spontaneous or found after the fluid has stood for several hours.
3. The presence of albumoses.
4. A greatly increased quantity of albumin.
5. No glycogen or, if present, a very small amount.
6. A striking capacity for hemolyzing the red-blood corpuscles of rabbit's blood.
7. The cell changes are nil, or, if any are present, they are slight.

When the disease has not progressed very far, the following findings:

1. Albumin present.
2. Albumoses present sometimes.
3. The yellow color very much diminished or entirely lacking.
4. No cell changes.
5. No hemolytic properties.
6. Glycogen content not diminished.

Sicard claims the above modifications are characteristic. In his cases, of twenty fluids examined ten were colorless and ten had the distinct yellow color; albumin was always present; in five cases the albumoses were found.

Mestrezat, however, questions whether these changes are not the typical changes of the syndrome of "coagulation massive et de xanthrochromie," of which twenty-one cases were collected by him from various sources.

"The syndrome of massive coagulation and xanthrochromie" is shown by typical cases or by so-called "cas frustes," which are variants from the usual type. The two main characteristics of the fluid of this syndrome are marked (quite often rapid) coagulability and the distinctly yellow color. The coagulation at times is so marked that the test-tube may be overturned without spilling any of the fluid.

Of the twenty-one cases collected, the first case was reported by Lepine; later cases by Froin[4] (1903) and Sicard;[5] then Babinski, Cestan and Ravant, Verhagen and Dustin,[6] and others. The primary underlying diseases and conditions were the following: meningomyelitis, tuberculous meningitis of cerebrospinal type, Landry's paralysis, compression of the cord by sarcoma, other neoplasms of the cord, pachymeningitis, and Pott's disease.

[4] Inflammations meningels avec reactions chromatique, fibrineuse, et cytologique du liquide Cephalo-Rachidien, Gaz. d. hôp., 1903, lxxvi, 1003; Syndrome de coagulation massive au cours d'une meningite, 1903, lxxxi, 1587.

[5] Syndrome de coagulation massive, de xanthochromie, et d'hemato-lymphocytose de liquide Cephalo-Rachidien, Gaz. de. hôp., 1908, lxxxi, 1431.

[6] Une sol-distant paralysis de Landry avec syndrome de Froin, Malde Pott cervical, Soc. d. Sci. med. et nat. (Bruxelles), 1909, lxvii, 169.

A resumé of the clinical signs and symptoms of the twenty-one cases is rather difficult to systematize and arrange. However, there is a generally habitual localization of the lesions at the level of the lumbar cord; most often paraplegias, spastic or flaccid, are found combined with sphincter disturbances, sensory disturbances, and sometimes painful subjective phenomena. In short, it is a process meningomyelitic in type, combined with a compression of the lower cord, which may arise from the above-mentioned factors. The symptoms then extend upward, as more of the cord is involved by the pathological changes, and a lumbar cul-de-sac is formed which is shut off from the remainder of the subarachnoid spaces.

Case XVII, reported by Verhagen and Dustin (1909), particularly concerns the subject of this article. Pseudoparalysis of Landry, paralytic and sensory disturbances quite well marked, beginning in the upper extremities, which finally involved the lower. The autopsy showed tuberculous caries of the fourth cervical vertebra making, by means of the pus, a medullary dorsal compression of the cord, and a fibrinous purulent exudate surounded the whole dorsolumbar portion. There was complete destruction of the lower cervical cord.

Thus this case of tuberculosis of the spine showed a cervical lesion compressing the cord with an exudate involving the whole dorsolumbar portion. This could easily give rise to the two factors which Mestrezat claims are necessary to the production of the typical findings of the spinal fluid of the syndrome, *i. e.*, a closed cavity of the lumbar region and a resulting stasis of the fluid. So by reason of the pathology the symptoms must perforce begin at a higher level and progress downward, being unlike the usual type of case. It seems that in this class the case now being reported should be placed.

A more detailed study of the fluid according to Mestrezat shows:

1. Coagulation immediate and spontaneous or after the fluid has remained standing several hours.
2. Yellow color, "xanthochromie."
3. Increased albumin content.
4. Presence of albumoses.
5. No changes in cell findings.
6. Sugar in variable amounts.
7. Endosmotic non-permeability of iodin and of the nitrates; exosmotic non-permeability of collargol and salicylate of soda.

PATHOGENESIS OF THESE ABNORMALITIES. Babinski (1903) explained the cause as hemorrhage; Sicard and Descomps (1908) gave the preference to a transudative origin of the fibrin and albumin found. These authors, without affirming the existence of a closed cavity in the lumbar region, insisted upon small meningeal pockets, whose walls played the role of transudation. On the contrary, Mestrezat insists upon two factors: (1) the existence of a closed cavity producing stasis of the fluid in which accumulated the elements issuing from the blood by way of transudation or hemorrhage; (2) an alteration, infectious or toxic in type, in the walls of the cavity, which so affects the vessels that a transudative process or small microscopic hemorrhages are created.

That the closed cavity is a factór may be shown physiologically by the non-permeability of the fluids to colloidal metals. Normally in ordinary meningitic cases (Mestrezat) colloidal metals injected in the spinal canal disappear rapidly. From the chemical point of view the existence of a cavity is evidenced by the facts that the albumins and fibrogens are increased, the spinal fluid is at a standstill and is not renewed, as a result of which no absorption of these elements takes place. Anatomically, by autopsies in six cases, a closed cavity has been demonstrated.

The "cas frustes" of the same author show deviations from the usual findings. These are due to the fact that the compression of the cord is not so forceful or that stasis of the fluid is not complete. Such cases lack one or more of the usual changes; thus the yellow color may be absent, or cell changes may be present and coagulability may be diminished.

The origin of xanthochromic, or the yellow color of the fluid in which no red-blood cells occur, is explained by Mestrezat as follows:

1. By serogenic means, as from transudative processes.
2. By hemolytic means, hemoglobin from entirely disorganized red-blood cells. Such fluids must be from a hemorrhage at least three days old.
3. By icteroid means, such as occurs in jaundice, when the spinal fluid shows a yellow color.

The diagnostic value of a fluid with a yellow color, therefore, is that it indicates icterus, minute meningeal hemorrhage, at least three days old, meningeal inflammation with intense congestion, or finally a symptom of the syndrome of massive coagulation and xanthochromic. Of course, this excludes the presence of actual hemorrhage into the canal, as evidenced by a yellow color and the discovery of red-blood corpuscles either macroscopically or microscopically, the "erythrochromie" of Mestrezat. This last is sometimes a manifestation of cerebral hemorrhage (Stewart[7]).

According to Nonne[8] (1913) the cause of xanthochromic has not yet been determined. He questions whether it is due to the passage of blood pigments through uninjured bloodvessel walls or is caused solely by many small hemorrhages. Schrützler claims the coloration comes not from blood pigments, but from color derivatives classed under the general group of luteins. The syndrome has been called by the Germans, "Kompressions Syndrome," Kleineberger, Rundfleisch, Erb, etc.

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[7] The Diagnosis of Nervous Diseases, London, 1914, chap. XIII.

[8] Deutsch. Ztschr. f. Nervenh., 1913, xlvii, 436.

In relation to the pathogenesis of the xanthochromie it is interesting to note the observations of Babes[9] (1914) in asystoliques (patients afflicted with a condition of the heart such that it is unable to make a complete systole, or a systole sufficient to expel the blood from the ventricles). In the examination of 10 cases, 8 showed a distinct color, 2 a less distinct; none of the patients had icterus, spinal fluids of all were negative to reaction for bile pigments. After examining these patients he then observed about 50 cases, divided into two classes. The first were local affections such as hernias, hemorrhoids, uterine fibromas. The second included more general maladies, as tuberculosis, pneumonia, acute appendicitis, etc. The first class showed no spinal fluids with yellow color, in the second it occurred in 5 cases (2 cases of pneumonia 2 of appendicitis and 1 of tumor albus, none showing any cord lesion, and 3 showing increase in albumin content). He explains the occurrence in the asystoliques as due to stagnation of the blood and to a transudation of the blood pigment through the vessel walls into the spinal fluid. In the second class of cases he thinks it is occasioned by minute capillary hemorrhages or to permeability of the vessels of the choroidal plexus and a consequent hemolysis of the red-blood cells. While these cases are irrelevant to the syndrome under discussion, they are of interest because of the occurrence of the xanthochromie, without the changes in coagulability, in cases showing no cord lesions or any pathological pressure on the cord.

It is my opinion that the case at present reported should be classed under the syndrome on account of the count of three cells to a field, the yellow color, and the presence of albumin, as shown both by the acetic acid and by the nitric acid test, and the slight coagulability. If the objection be raised that the coagulation was not "massive," it can at least be classed as one of the "cas frustes." As additional evidence, there are the Roentgen-ray plates, showing an evident caries of the third and fourth lumbar vertebræ, and the paralysis and sensory disturbances affecting chiefly the right side of the body. No localization pointing to the lumbar region occurs, but it does resemble the case of cervical caries of Verhogen and Dustin. It is to be regretted that no necropsy was permitted. It is a fair assumption, however, that the process in the cervical vertebræ produced a compression of the cord, and as a result the paralysis and sensory disturbances. In addition a closed cavity, a cul-de-sac, occurred and a consequent stasis of the cerebrospinal fluid. In spite of the fact that the Roentgen-rays showed most probably some tuberculous involvement of the lungs, from any

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[9] La xanthochromie du liquide cephalo-rachidien, dans d'autres maladies que les hemorragies cerebrales, les affections du nevraxes et l'ictere. Compt. rend. Soc. de biol., 1914, lxxvi, 671: La xanthochromie du liquide cephalo-rachidien chez les asystoliques, Rend. Soc. de biol., 1914, lxxvi, 313.